The Rare Case of Mucosa Associated Lymphoid Tissue Lymphoma in Ileum

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Abstract
The body has mucosa-associated lymphoid tissue (MALT), with the gastrointestinal (GI) tract having the greatest amount of it. Lymphoma may form when the cell growth in this tissue is aberrant. The small intestine is a common extranodal site of lymphoma, which is a systemic illness. Additionally, it has been proposed that MALT lymphomas (MALTomas) arise as a result of chronic and persistent immunological activation, whether of an autoimmune or infectious type. MALT lymphomas that develop in the duodenum are typically thought to be unrelated to Helicobacter pylori infection. However, some examples show that lymphoma regressed when H. pylori was removed.

Keywords: and cd21+, cd35+, cd20+, igm+, igd, marginal zone b cells, h. pylori infection., mucosa-associated lymphoid tissue (malt) lymphoma

Introduction
Mucosa-associated lymphoid tissue (MALT) lymphoma was described first by Isaacson and Wright in 1983. They found that peripheral lymph nodes and mucosa-associated lymphoid tissue share more histological characteristics with primary low-grade gastric B cell lymphomas and immunoproliferative intestinal illness. MALT lymphomas can develop at a variety of extranodal sites, such as the stomach (70%), the lung (14%), the ocular adnexa (12%), the thyroid (4%), and the small intestine [1]. MALT is an extranodal marginal zone B-cell lymphoma [2]. It is the cancerous multiplication of B cells of lymphoid tissue in the marginal area [3]. Morphologically it is heterogeneous small B cells which include marginal cells. The cells resemble small lymphocytes, centroblast-like cells, scattered immunoblasts, and monocyteid cells. Also, they include invaded lymphoma cells throughout the epithelium with the multiplication of plasma cells in the lamina propria of the mucosa [4]. MALT symptoms differ based on the site where the organs are affected because it is a localized condition. In fewer than 5% of instances, it has non-specific symptoms, including malaise, weight loss, low-grade fever, and abdominal pain [5].

Case Presentation
A 38-year-old male patient was apparently alright two days back, then he developed sudden onset of pain in the abdomen, which was colicky in nature, non-radiating, moderate to severe and gradually progressive in intensity, associated with one episode of non-bilious non-projectile vomiting with food and water content. He has not passed stools for the past two days. There is no history of fever, cough, or burning micturition. There was no significant past medical or surgical history. He occasionally drank about 60ml of alcohol once a month. The general physical and systemic examinations were normal except for tenderness in the right iliac fossa. Contrast-enhanced computed tomography (CECT) abdomen and pelvis was done which was suggestive of telescopic appearance in the ileal segment intussusception.

The patient underwent resection and anastomosis of the ileal segment (Figure 1) and (Figure 2).
FIGURE 1: Shows intraoperative picture immediately post opening up of the abdomen.

The black arrow shows intussusception with the classical telescopic appearance of the ileal segment.
FIGURE 2: Nodular growth noted over the ileal segment.

The black arrow shows the site of MALT acting as a lead point for intussusception.

MALT: Mucosa-associated lymphoid tissue

Postoperatively the specimen was examined and the gross appearance of the resected ileal segment was suggestive of a growth over which intussusception was present (Figure 3).
FIGURE 3: Postoperative photo of the resected specimen.

The specimen was sent for histopathological examination and was suggestive of non-Hodgkin’s lymphoma (Figure 4) and (Figure 5).
**FIGURE 4:** Non-Hodgkin's lymphoma diffuse pattern MALT type MALToma.

At 40x magnification. Hematoxylin and eosin staining

MALT: Mucosa-associated lymphoid tissue
Postoperative period was uneventful and was discharged with a six monthly follow-up date.

Discussion

The body has mucosa-associated lymphoid tissue, with the gastrointestinal (GI) tract having the greatest amount of it. Lymphoma may form when the cell growth in this tissue is aberrant. MALT lymphoma of the GI tract makes up 50% of cases that are present. The small intestine, the stomach, and the colon are the next most frequent sites where MALToma can occur. Regarding the location of small intestine lymphomas specifically, they account for up to 40% of primary GI lymphomas and 25% of primary malignancies at this level. The small intestine is a common extra-nodular site of lymphoma, which is a systemic illness [6]. The ileum and cecum in the intestine are the most affected areas, likely as a result of the plentiful lymphoid tissue there. Multiple localizations are also seen in 5%-15% of cases [7]. Strong correlations have been found between *Helicobacter pylori* infection and gastric MALT lymphomas. This is the most prevalent infectious agent connected to worldwide cancers during an *H. pylori* infection. Three chromosomal translocations- t(11;18) (q21;921), t(1;14)(p22;q32), and t(14;18)(q32;q21)-help turn healthy B cells into cancerous clones. Nuclear factor kappa B (NF-B), which is implicated in immunity, inflammatory processes, and apoptosis, is stimulated as a result of these translocations [8]. Additionally, it has been proposed that MALTomas arise as a result of chronic and persistent immunological activation, whether of an autoimmune or infectious type [9]. A primary gastrointestinal lymphoma must meet five requirements outlined by Dawson, including the lack of peripheral lymphadenopathy at the time of presentation, the absence of enlarged mediastinal lymph nodes, a normal total and differential white blood cell count, the predominance of a bowel lesion at the time of laparotomy with only nearby lymph nodes clearly affected, and the absence of lymphomatous involvement of the liver and spleen [10]. The invasion of the marginal zone and the diffuse dissemination of
the tumor into the surrounding tissue are the histopathological characteristics of MALT lymphoma. MALT lymphoma cells exhibit the same cytological and immunophenotypic characteristics as marginal zone B cells (IgD, IgM+,CD20+, CD35+, and CD21+) [1]. Multiple tiny erosions, nodular elevations, and diffuse erythema in the duodenal bulb are the symptoms of duodenal MALT lymphomas. According to specific descriptions, the symptoms consist of several ulcer crater without stomach involvement, each having nodular and erythematous edges.

MALT lymphomas that develop in the duodenum are typically thought to be unrelated to H. pylori infection. However, some examples show that lymphoma regressed when H. pylori were removed. An unusual discovery is MALT lymphomas of the ileum. According to early descriptions, the endoscopic pattern consists of many reddish or white masses with a polished and smooth mucosa look. Five patterns were distinguished in the first classification of ileal lymphoma endoscopic features: mucosal fold thickening alone, nodular pattern, infiltrating pattern, ulcerative pattern, and mosaic pattern [7]. Even though it is rare, a possible complication is when a proximal section of the intestine telescopes into the lumen of the neighbouring section during intussusception. Intussusception in adults is incredibly uncommon and typically only occurs in younger populations. Adult symptoms can be very different. As nonspecific symptoms of adult intussusception, bleeding, nausea, changes in bowel habits, vomiting, and constipation with abdominal swelling has been noted [2]. The development of capsule endoscopy and the double balloon technique of push-and-pull enteroscopy has improved the diagnosis of small intestinal lymphomas. Ileal MALT lymphoma is anecdotical, and only a handful of occurrences have been reported in the literature. The majority of cases show a lack of H. pylori infection, leaving the cause of this condition unexplained. Nearly half of patients present with a single tumour that resembles a carcinoma in physical appearance; multiple masses or lymphomatoid polyposis are less common [7]. By performing a biopsy on the afflicted area and morphologic, immunophenotypic, and genetic studies on the biopsy specimen, a reliable diagnosis of MALToma can be obtained [11]. For the detection of MALTomas, positron emission tomography–computed tomography, or positron emission tomography/computed tomography (PET-CT), is more effective. In one study, 42% of early MALTomas were found using PET-CT [12].

The most sensitive procedures are endoscopic submucosal resection or ultrasound-guided endoscopic fine-needle aspiration biopsy [3]. Nongastric MALT lymphoma treatment after diagnosis is identical to that for other non-Hodgkin lymphomas. A thorough physical workup, a laboratory assessment that includes a full blood count with differentials, an all-inclusive metabolic panel, and a quantification of lactate dehydrogenase, as well as a CECT of the pelvis, chest, and abdomen, should all be included in the workup [2]. Nearly 90% of patients with stomach MALT lymphoma survive for 10 years, and about 70% remain disease-free [1].

Conclusions
The third most prevalent non-lymphoma Hodgkin’s subtype is MALToma. Since MALToma of the ileum is uncommon, treating H. pylori, which is a major factor in the formation of MALToma, should be the first line of treatment. It is difficult to make a quick clinical diagnosis because the diagnosis is only made intraoperatively for intestinal blockage. The incorporation of both chemotherapy and surgery is thought to be superior to other approaches. In the case described above the ileal segment was resected and anastomosis was done. The incidence rate, as well as the proper diagnostic and treatment procedure for intestinal MALToma, is not standardized in previous studies and literature which proves to be a major problem.

Additional Information
Disclosures
Human subjects: Consent was obtained or waived by all participants in this study. Conflicts of interest: In compliance with the ICMJE uniform disclosure form, all authors declare the following: Payment/services info: All authors have declared that no financial support was received from any organization for the submitted work. Financial relationships: All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. Other relationships: All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

References